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# FIBRO-OSSEOUS LESIONS OF THE HEAD AND NECK

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**In recent years the term "fibro-osseous lesions" has gained wide acceptance as a general designation for certain pathological processes of the craniofacial bones. These lesions comprise a challenging group of pathologic conditions that cause difficulty in classification and treatment. Common to all of these pathologic entities is the replacement of normal bone architecture by a benign fibrous tissue composed of fibroblasts and collagen and which contains varying amounts of mineralized material. Categorization of the fibro-osseous lesions is dependent upon correlation of the patient's history, clinical findings, radiographic criteria, and histopathologic determination. This article discusses two of the most commonly confused benign fibro-osseous lesions: fibrous dysplasia and ossifying fibroma.**

Fibrous dysplasia, first described by Lichtenstein in 1938, and then by Lichtenstein and Jaffe in 1942, was considered distinct from other fibro-osseous lesions.[1,2] More recently however, the term "benign fibro-osseous lesions" has been used to describe fibrous dysplasia as well as other pathologic entities. Fibrous dysplasia is a fibrous-osseous lesion of unknown etiology that results from an abnormality of development of bone-forming mesenchyme in which an area of normal bone is replaced by fibrous tissue. This fibrous tissue undergoes metaplasia which results in increased bone mass. The disease has three clinical forms: (1) monostotic (isolated to one bone), (2) polyostotic (affecting two or more bones), and (3) Albright's syndrome (polyostotic form associated with abnormal skin pigmentation, precocious puberty, and other nonskeletal manifestations). The proportion of monostotic to polyostotic forms is 40.

## CLINICAL PRESENTATION

Dysplasia usually becomes manifest in the second decade of life, grows slowly, and then stabilizes in adult life. Occasionally the disease reactivates during pregnancy and, in some instances, it may continue to grow slowly throughout life. There is no known predisposing underlying disease state and the disease is twice as common in females. The craniofacial bone most commonly involved in the monostotic form is the maxilla[3] with the mandible affected second in frequency. The calvarium is infrequently involved as are the sphenoid[4] and ethmoid sinuses.[5] In the polyostotic form, bones of the lower limbs are involved more commonly. A small percentage of patients with polyostotic fibrous dysplasia have a condition referred to as Albright's syndrome. This syndrome is characterized by cafe au lait pigmentation of the skin, precocious puberty in females, and other extraskeletal abnormali- ▶

ties. The frequency of head and neck involvement in Albright's syndrome is not predictable, but when involved, the base of the skull and occiput are most severely affected.

Fibrous dysplasia causes a slow but progressive facial asymmetry that is usually painless. The proliferation of fibrous dysplasia causes displacement of various structures in the facial skeleton and is occasionally diagnosed as a developmental anomaly.[6] The lesions can attain considerable size with 60% measuring 4 cm to 8 cm in greatest dimension.[7] Teeth are frequently present in lesions affecting the alveolus and, although malposition may occur, marked loosening is not usual.

## RADIOLOGICAL FINDINGS

Radiological appearance of fibrous dysplasia is variable depending upon the amount and distribution of osteoid matrix in the lesion. In one series of 46 consecutive cases of fibrous dysplasia of the craniofacial skeleton, the radiographic diagnosis in each case was found to correlate accurately with pathologic analysis.[8] The classic radiographic appearance of fibrous dysplasia is described as an "orange peel" or "ground-glass" radiopacity that is not well circumscribed. Occasionally, areas of radiolucency and cortical thickening may be observed. CT imaging is most valuable for the diagnosis and delineation of temporal bone lesions; expansile growth, thinning of the surrounding cortical bone, and displacement rather than destruction of adjacent structures are characteristic features. The otic capsule is usually spared and may appear to float within the lesion.[9]

Physicians should not rely solely on radiographic analysis for the diagnosis of fibrous dysplasia, but should also obtain histologic confirmation. Included in the differential diagnosis of lesions which may appear similar to fibrous dysplasia on radiologic imaging are Paget's disease of bone, hemangiomas of bone, giant cell tumors, aneurysmal bone cysts, and ossifying fibroma.

## HISTOPATHOLOGY

The histopathologic appearance of fibrous dysplasia is distinct from that of ossifying fibroma. The normal components of the marrow are replaced by whorled spindle cells and irregular woven bone trabeculae. These trabeculae of osteoid tissue are arranged in a completely meaningless arrangement that forms V or W shapes resembling Chinese characters. The monostotic and polyostotic forms are indistinguishable histologically. Lack of osteoblastic rimming is a recognized criterion for the diagnosis of fibrous dysplasia. Deep biopsy is essential to demonstrate the essential defect in bone maturation and probably explains the difficulties in differentiating between ossifying fibroma and fibrous dysplasia on histologic grounds alone.

## TREATMENT

Management begins with establishing the diagnosis on clinical, radiographic, and histopathologic grounds. Small lesions of the jaws that cause only minor intraoral contour distortions or that are discovered serendipitously through routine radiographic examination probably require no definitive care after biopsy diagnosis. In patients in whom there is significant cosmetic distortion or functional disturbance, such as malocclusion, surgical treatment requires careful timing. Surgery is essentially a sculpturing maneuver to provide the patient better social acceptance or a functional masticatory apparatus. Since surgery is rarely able to eradicate the entire disease process, it is preferable to delay surgery until the growth phase of the patient has slowed. Therefore, it is unwise to operate on the adolescent because of the potential for continued progression of the dysplastic process after surgery. Some authors have reported an acceleration of the disease after surgery, seemingly provoked by the surgery itself. In patients with significant deformities, however, social or functional considerations mandate early debulking, with the anticipation that a repeat procedure will be necessary at a later date. Radiation treatment is contraindicated in the management of fibro-osseous lesions.

## OSSIFYING FIBROMA

Ossifying fibroma, also known as cementifying fibroma and cemento-ossifying fibroma, was first described by Mandel[10] in 1872 as occurring in a 35-year-old woman with a tumor of the mandible which had been present for 25 years. In the following years there was some confusion regarding the various fibro-osseous lesions, especially ossifying fibroma versus fibrous dysplasia. However, present opinion is that

ossifying fibromata are a distinct entity, differing from monostotic dysplasia, and distinguished by distinct clinical, radiological, and histological criteria.[11] A possible origin from both the mesenchyme of bone and from the periodontal membrane has been proposed for this fibroma.

*Clinical Presentation.* Some consistent clinical differences exist between ossifying fibroma and fibrous dysplasia.[12] Although the ossifying fibroma occasionally occurs in the long bones, it is predominantly confined to the head and neck and usually develops as a single entity in the jaw. Although most of these lesions present as sclerotic lesions in the premolar area of the mandible, posterior maxillary involvement has been reported. The lesion can occur in any age group, but the third and fourth decades of life are the most common periods of occurrence. When the ossifying fibroma appears in childhood, it usually grows rapidly. It forms a localized, nontender swelling with displacement of the teeth as a common clinical finding. Large lesions show thinning of the cortical plate and bowing of the inferior border of the mandible.

*Radiological Findings.* On radiographic examination, ossifying fibroma produces a well-circumscribed area of radiolucency often with a sclerotic margin and small radiopaque masses within the lesion. To a lesser degree than fibrous dysplasia, the ossifying fibroma may blend into normal bone, causing some degree of difficulty in distinguishing it from normal osseous structures. The most significant feature that distinguishes the ossifying fibroma from fibrous dysplasia is the circumscribed nature of the ossifying fibroma.

*Histopathology.* On histologic examination, ossifying fibroma possesses cellular fibroma connective tissue stroma containing rims of osteoblasts and trabeculae of mature (lamellar) bone. Although the osseous component is generally described as mature, the central portions may be woven bone with lamellar bone at the periphery; complete bone maturation is seldom seen.

*Treatment.* Since this process is benign and relatively discrete, lesions are surgically well delineated from adjacent bone, and enucleation is the treatment of choice. An intraoral approach is usually employed in surgical removal of these lesions, and recurrence after surgical removal is rare.

## CONCLUSION

We have attempted to compare and contrast the two main fibro-osseous lesions, fibrous dysplasia and ossifying fibroma. An attempt to definitively diagnose any fibro-osseous lesion of the craniomaxillofacial region by clinical, radiographic, or histologic features alone is very dangerous. A careful history must be obtained and a careful physical examination must be performed on all patients presenting with bony lesions of the head and neck. The differential diagnosis of fibrous dysplasia from ossifying fibroma rests on a radiological criterion after the histopathologist has verified the fibro-osseous nature of the lesion. The clinical and radiographic findings, more than the results of the biopsy, will dictate to the surgeon whether or not his task is one of recontouring a generally nonresectable lesion of fibrous dysplasia (unless the lesion is small) or whether he is obliged to resect completely a generally well-outlined ossifying fibroma in toto, with the goal of total eradication.

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